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RECENT studies of the artefacts of the nervous system by Van Gieson, some of the results of which were presented at a meeting of the New York Neurological Society in June, 1892, and subsequently embodied in a series of articles in the *New York Medical Journal*, place in an entirely different light many of the heretofore recorded cases of congenital abnormality of the spinal cord. Van Gieson, after an exhaustive review of the cases of malformation of the cord reported prior to 1892, together with an experimental study of the subject, including the examination of a large number of cords unwittingly injured during removal, and others in which it was attempted by intentional bruising and rough treatment to reproduce some of the appearances previously regarded as of ante-mortem origin, reaches the conclusion that of the thirty-one cases of supposedly congenital malformation recorded from 1878 to 1892, only six are instances of true developmental defect; the remaining twenty-five, including cases of heterotopia of the gray and white substances, and several instances of most remarkable doubling of the cord, reported by Fürstner, Zacher, Drummond, Schiefferdecker, Feist, Kronthal, and others, being nothing more than topographic alterations in the nervous tissues produced by bruises and other injuries inflicted during unskilled or careless re-



moval of the cord from its bony canal, or by rough handling after its extraction.

When it is considered that the literature of these pseudo-malformations had attained to quite respectable proportions, and that the misconception of their true nature, passing unchallenged for many years, had already given rise to erroneous conclusions as to the frequency of spinal-cord malformations in general, and of their influence in the causation of disease, the value of Van Gieson's work may be appreciated.

The case which is made the subject of this paper offers an apt illustration of the facility with which the most bizarre and misleading appearances may be produced by unintentional injury of the cord during post-mortem examination; and but for Van Gieson's rational explanation of the real character of the apparent anomalies noted, it would in all probability have been added to the twenty-five instances of spurious "congenital malformation" already recorded.

The patient was an epileptic imbecile girl, subject to convulsions from early childhood, small, and misshapen; the cranium, face, and body were asymmetric, the right side the smaller; there was a slight lateral curvature of the vertebral column, with marked displacement of the spines of the eleventh and twelfth dorsal vertebræ. The convulsive movements usually began and were more severe upon the right (the smaller) side. The tendon-reflexes were exaggerated, the more markedly so upon the right side; a distinct ankle-clonus existed on this side. The gait was shambling and unsteady, but not characteristic. The patient was irritable, possessed a limited vocabulary and very slight intelligence. She died in her twenty-sixth year, of tuberculosis of the lungs.

The autopsy, held five hours after death, showed extensive and extreme sclerosis of the first and second temporal convolutions and of the cornu ammonis of the left hemisphere; the *right* lateral lobe of the cerebellum was

smaller than the left, and its posterior portion shrunken and sclerosed. The discovery of these lesions led to a more than ordinarily careful removal of the spinal cord, this being accomplished without appreciable injury to the organ, save at the point of displacement of the eleventh and twelfth vertebræ, where some difficulty was experienced in getting through the laminæ, and a diagonal gash in the dura and enclosed cord was inflicted by the chisel. After the cord, enclosed in its membranes, was taken out, it was observed that at the dorso-lumbar junction the cord seemed remarkably small, and this fact was noted in the written report of the autopsy. This abnormally contracted portion extended from the point of injury upward for about one and one-fourth inches; the small cord could be distinctly felt through the relaxed dura; at the upper extremity of the contracted segment the cord became unduly large, and the dura seeming tensely distended by its contents; this swelling gradually diminished above, and within a distance of one and one-half inches the cord had regained its natural size. It was conjectured that this abnormality might in some way be associated with the deformity of the spinal column and the symptoms of a spinal lesion noted during life. The tissue, seeming soft, the cord was placed in Müller's fluid for hardening without further examination or handling, a few transverse cuts being previously made through cord and dura, but leaving untouched the seat of the supposed malformation. The process of hardening was completed in alcohol, and the cord then subjected to careful inspection before dividing it into convenient bits for section-cutting.

Transverse cuts through the dura and the cord at the point of greatest apparent hypertrophy showed the seemingly normal and uninjured dorsal cord crowded to one side, and the entire space between the cord and the dura filled by a crescent-shaped mass of material, softer and paler than the cord, with a somewhat mottled surface.

The cord and the outlying mass were apparently enveloped in the same unbroken arachno-pial sheath. Sections above and below this point discovered the extraneous crescent to thin away—gradually above, abruptly below, where it terminated in the small contracted portion previously mentioned. A cross cut through this contracted area disclosed the appearance of a cord shrunk to one-third its normal size, and destitute of gray matter; below, at the point where the cord resumed its normal dimensions, the wound made by the chisel, with some bruising and extrusion of the nervous tissue, was noted; over the remainder of the contracted part, as at the swelling above, the arachno-pial covering seemed uninjured. A transverse section at the junction of the swollen segment with the seemingly atrophic segment showed a considerable distortion of the gray horns of the cord and some pale splotches in the white tissue; several similar but smaller whitish patches were noted in the columns of Goll in the upper portion of the cervical enlargement. Throughout the remainder of its length the cord seemed to the naked eye to be of normal character, excepting for the disintegration observed at the point of injury in the upper lumbar region, previously referred to.

Microscopic examination of a section prepared from tissue taken from the abnormally enlarged segment resulted in the unexpected discovery that the extraneous crescent was composed of nervous tissue, gray and white, the gray arranged so as to present a rude imitation of the normal disposition of the gray matter of the spinal cord, the white showing irregular bundles of nerve-fibers interspersed between masses of apparently unorganized myelinic material; in brief, an instance of partial duplication of the spinal cord had been discovered. Some regret was felt that a cord exhibiting so interesting and unusual an anomaly should have been injured just at the seat of the malformation, and a detailed study of the organ was undertaken. The cord was cut into half-inch

pieces and sections made representing practically the entire length of the cord; at the swollen and contracted parts special care was exercised, and sections in series to the number of many hundreds were made and examined.

Beginning above, the first abnormality discovered was in the upper portion of the cervical enlargement, where, in the columns of Goll, the irregular pale spots had been noted; these the microscope showed to be due to an apparently anomalous course of some of the bundles of

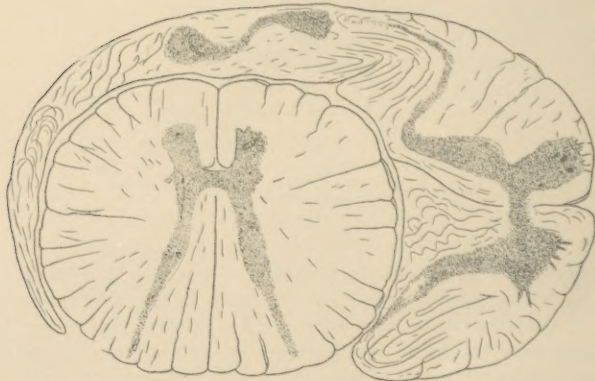
FIG. 1.



nerve-fibers; the fibers, instead of running longitudinally and showing their cut ends only in a cross-section of the cord, pursued a more or less horizontal course, and were traceable for considerable distances across the sections; some fibers pursued a peculiar gyral or corkscrew like course; some ran in straight bundles, the fibers lying parallel; some exhibited an extremely irregular arrangement. The abnormality was entirely confined to the columns of Goll, and to the left anterior pyramidal tract (Fig. 1). From this point to the beginning of the abnormal swelling in the lower dorsal region

no lesions or abnormalities meriting notice were detected. The sections then bring into view the first evidence of the outlying crescent of nerve-tissue in two separate masses of nerve-material, composed in part of bundles of medullated nerve-fibers, in part of apparently unformed myelinic material, staining deeply by the Weigert hematoxylin method. A little lower down these two masses become united, and two small patches of gray substance make their appearance. A short distance below this point the gray and white masses assume somewhat the appearance of a much-distorted anterior half of a spinal cord, which likeness becomes more pronounced when the point of greatest apparent hypertrophy is reached (Fig. 2). Here the supernumerary cord is well shown,

FIG. 2.



although there is much distortion of the gray horns and many abnormalities of arrangement in the white tissue. The gray horns are larger than the gray horns of that portion of the dorsal cord around which the heterogeneous tissue has disposed itself, and the surface-area of

a transverse section of the accessory cord is somewhat greater than that of the dorsal cord. Below (Fig. 2) the abnormal crescent diminishes rapidly in thickness, and the cord shows the first indications of intrinsic malformation; the posterior gray horns become unduly ap-

FIG. 3.



proximated, and some distortion of the white substance, notably in the posterior columns and on each side of the anterior median fissure, appears (Fig. 3). Immediately below this point the distortion of the gray and white substances of the cord becomes extreme; large bundles of fibers run diagonally across the section, pass through the ruptured pia, and apparently become continuous with the extraneous crescent above (Fig. 4). The cord then becomes contracted, and a transverse section shows an appearance but little resembling a spinal cord; there is simply a mass of irregularly disposed white fibers and myelinic material contained in a collapsed and shrunken arachno-pia; at the lower extremity of this shrunken part the wound inflicted by the chisel in the dura, pia, and cord is situated, and some of the cord-substance is

extruded through the gash; from a point immediately below the wound to the extremity of the cord no lesion is discovered.

The features of noteworthy interest, in addition to the topographic changes illustrated in the diagrams and in the foregoing description are these: The extraneous crescent is continuous below with the mass of nerve-tissue passing out of the cord, shown in Fig. 4; and here the pia is ruptured; the extruded material and the entire mass of the outlying crescent throughout its length occupies the "sub-arachnoid space"; the arachnoid is no-

FIG. 4.



where broken, save at the point of chisel-injury, some distance below this. The heterogeneous crescent gives origin to no nerve-roots, and no bloodvessels can be traced into it.

Many hours were spent in an attempt to solve the problem of the development of this accessory cord in its peculiar situation, and of the abnormal contraction and absence of gray matter in the cord immediately below this. In looking up the literature of the subject, reference to partial doublings, somewhat resembling this case, was found in one or two of the text-books and in the

writings of several German authors. The results of injury in the upper lumbar region were recognized, but it was not deemed possible that so remarkable a duplication of the cord could be produced by injury of any kind or degree. Being, however, unable to arrive at any intelligible solution of the puzzle, the whole matter was put aside until recently, when, through the courtesy of Dr. Welch, of the Johns Hopkins University, to whom a section of the cord was submitted, Van Gieson's monograph was brought to notice.

In the light of Van Gieson's researches the case is made plain. The entire abnormality is a *bruise artefact*.

The spinal cord is of especially difficult access at the dorso-lumbar junction, and it is in exactly this locality that most of the reported duplications of the cord have been situated. In the case in hand the displacement of the vertebræ made the severance of their laminæ a matter of peculiar difficulty; the chisel employed in effecting this by some mishap cut through the dura and into the pia-arachnoid and the cord itself. The spinal dura is a tough and by no means easily lacerated membrane, so that a cut through it with a chisel not particularly sharp, presupposes the application of considerable force; in this instance the cord was probably severely bruised and a powerful squeeze applied to it before the dura gave way. This violence was sufficient to detach a fragment of cord-tissue, and, rupturing the pia, forced the loosened segment upward into the sub-arachnoid space, Fig. 4 representing the point of exit. The distortions and topographic anomalies in the cord, illustrated in Fig. 3, are doubtless results of the same violence which caused the invagination. The lesser abnormalities, shown in Fig. 1, are due to an appreciated small bruise at this point (upper portion of the cervical enlargement). The invaginated crescent, of course, shows much disintegration of its structural elements and alterations in the relations of

its gray and white substances ; it is torn loose from its nerve-roots and nutrient vessels. The abundant formless myelinic material mentioned as taking a deep Weigert stain is derived from the crushed and broken myelin-sheaths of the white nerve-fibers. The cord, apparently shrunk to one-third its normal size, is the column of white substance remaining after the separation and upward invagination of the portion which goes to form the distorted crescent.

During the past several years probably twenty-five or more spinal cords have been removed in our autopsy-room in about the same manner as the one under consideration, and sections from them studied with greater or less care. In no one of these cases, however, has any appearance at all resembling those above sketched been discovered. Several of the cords have been injured in removal, but the results of injury have always been easily recognizable as such ; and the case immediately under consideration is the only one in which any such deceptive duplication has been unearthed.

It is a matter of some practical importance to workers in the pathology of the nervous system that the fact be recognized that, however rarely, such remarkable doublings as the one here reported do occur as a result of unintentional violence to the cord, and perchance when least expected.

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